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## **Pediatric palliative care-when quality of life becomes the main focus of treatment**

Bergstraesser, Eva

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# Pediatric palliative care—when quality of life becomes the main focus of treatment

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**Abstract** Pediatric palliative care (PPC) focuses on children and adolescents with life-limiting diseases. It may be initiated at various points of the disease trajectory, if possible early enough to support living with the best possible quality of life despite a limited lifespan. From birth to adolescence, children with a broad spectrum of diseases may benefit from PPC. Since 50% of deaths in childhood occur within the first year of life, PPC is just as relevant to neonatology. Causes of death in the neonate and young infant are due to perinatal conditions such as preterm birth and congenital disorders and syndromes; in older children, external causes, such as traumatic injuries, outweigh disease-related causes of death. PPC may last from a few hours or days for neonates to many years for children with complex chronic conditions. For neonates, PPC often has the character of end-of-life (EOL) care followed by bereavement care for the family. For older children, PPC can clearly be differentiated from EOL care; its indications include progress or deterioration of disease, marked instability of the child's condition, increase in the need for technical or medical support, increase in suffering, or failure of treatment. If a child's need for palliative care is established, useless and potentially harmful treatments may be withheld and informed choices can be made about treatment, care, and the remaining life of the child. **Conclusion** This review aims to provide knowledge for clinicians who care for children and adolescents at risk of dying from their disease. PPC can improve the child's remaining lifetime by focusing on quality of life and goals that are defined by the child and his or her family.

**Keywords** Pediatric palliative care · End-of-life care · Children · Definition of palliative care in children

## Abbreviations

PPC pediatric palliative care  
EOL end-of-life  
CCC complex chronic condition  
ACT Association for Children's Palliative Care

## Introduction

Despite great advances in medicine, children<sup>1</sup> suffering from life-limiting illnesses and their families may still be confronted with the unnatural fact of a shortened lifespan. Pediatric palliative care (PPC), a relatively young discipline, focuses on this patient group and strives to ease suffering and enhance quality of life. Many readers may think that this is an inherent part of medicine and does not need specialization. I agree to some point, however, there might be a specific role for specialized physicians in PPC, as in other specialties. This review article aims to provide an overview of current pediatric palliative care.

## Background

### Historical aspects

The relief of symptoms and suffering has always been a component of a physician's duties, and, in the very beginnings, it frequently was the only goal of medicine. However, with exciting advances in medicine, the patients' suffering

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<sup>1</sup> "Child" also includes the adolescent but not the neonate.

has increasingly been neglected. This is probably most striking in oncology and may, therefore, explain why palliative care, including PPC, has its main sources there [6, 40].

PPC began to rouse scientific interest in the beginning of the current millennium. This is underlined by two sentinel articles; one published in *Pediatrics* [1] by the American Academy of Pediatrics (AAP) and the other in *The New England Journal of Medicine* [54] by Wolfe et al. from the Dana-Farber Cancer Institute. The AAP emphasized in its recommendations [1] that “Program development in pediatric palliative care, along with community outreach and public education, must be a priority of tertiary care centers serving children.” Wolfe et al. used the example of the suffering of children dying from cancer whose problems had, by then, triggered scientific curiosity. Meanwhile, symptom management has improved at least for children dying from cancer [55].

Prior to the scientific approach, palliative care as a discipline originated in the hospice movement for adults in the UK of the 1960s with the ambitious activities of Cicely Saunders and the opening of St. Christopher’s Hospice [10]; for children, the movement started in the 1980s in the UK, Canada, and the USA [5, 11, 36].

#### Definition of pediatric palliative care and associated terms

There is a high variability in current practices and attitudes regarding palliative care for children with life-limiting diseases [49]. One reason for this may be the philosophical and holistic nature of the definitions of palliative care that do not provide practical aspects of care. Such definitions, including those on PPC, are based on the definition of the World Health Organization [57]:

Palliative care is an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual. Palliative care:

- provides relief from pain and other distressing symptoms;
- affirms life and regards dying as a normal process;
- intends neither to hasten nor postpone death;
- integrates the psychological and spiritual aspects of patient care;
- offers a support system to help patients live as actively as possible until death;
- offers a support system to help the family cope during the patient’s illness and to deal with their own bereavement;
- uses a team approach to address the needs of patients and their families, including bereavement counseling, if indicated;

- will enhance quality of life, and may also positively influence the course of an illness;
- is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications.

Palliative care for children represents a special field of palliative care, even though it is closely related to adult palliative care. In Europe, the definition by the Association for Children’s Palliative Care (ACT) is the most common [19]:

Palliative care for children and young people with life-limiting conditions is as an active and total approach to care, from the point of diagnosis or recognition, throughout the child’s life, death and beyond. It embraces physical, emotional, social and spiritual elements and focuses on enhancement of quality of life for the child/young person and support for the family. It includes the management of distressing symptoms, provision of short breaks and care through death and bereavement.

In the context of PPC, two additional terms need to be defined, i.e., “life-threatening” and “life-limiting” disease. The former describes a disease for which a cure is realistic but may fail, and the latter describes a disorder for which there is no hope of cure. PPC has its focus on life-limiting diseases and thus on a limited lifespan even if it may be applied earlier on, or in serious illnesses with prognostic uncertainty.

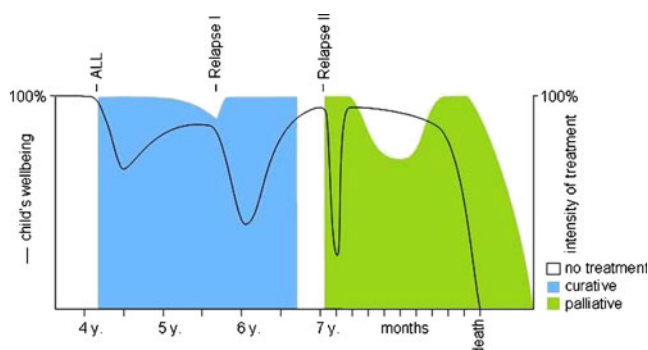
In contrast to palliative care in adults, prolonging life is a crucial aim in PPC [31]. Furthermore, it is emphasized that end-of-life (EOL) care or terminal care is part of palliative care; however, it encompasses the phase of dying. Laypeople but also health professionals still misunderstand palliative care to be dedicated to the end of life. This may result in destabilization and defense particularly in those who could benefit from a palliative care approach. In addition, the process of redefining treatment goals may also be inhibited by a widespread phrase used by physician: “We are not there, yet.”

Why is it so important to define a palliative care status for a child and the family? Several answers emerge: (1) to prevent useless treatments which may even be harmful, (2) to allow the patient and the family to make informed choices about treatment and care, (3) to allow for autonomy by defining goals for the remaining life span, (4) to reassure the severely ill child or adolescent that his or her life has meaning and purpose and he or she is loved and will always

be remembered, (5) to help prepare for imminent death; this may include the wish of a child to say good-bye or to leave meaningful thoughts or things for those who are left behind, and (6) to allow for the planning of death, including location and setting (Vignette 1 demonstrates these aspects of PPC including bereavement care.) (Fig. 1).

**Vignette 1** Second relapse of leukemia (ACT group 1 [19])

Urs is a 7-year-old boy when the second relapse of an acute lymphoblastic leukemia restricted to the central nervous system (CNS) is diagnosed (first diagnosis at the age of 4, first relapse 20 months later). He complains of severe headaches and vomiting. His parents clearly vote not to bother him too much as he and the whole family suffered a lot during the former two treatments. Fortunately, Urs responds well to a CNS-directed treatment and repeated pressure release through an Omayra reservoir which has been implanted to ease treatment. His cerebral fluid can be cleared of blasts and his condition becomes nearly normal again. He plays football and tennis outside and his first school day is planned. Five months later, the whole family takes a vacation. The leukemia has disseminated at this point, which is why provisions are taken for a potential transfusion of platelets on the Canary Islands. However, it turns out that Urs does not need a transfusion during this vacation, and the family enjoys what, very likely, is their last holiday together. Following the holiday, Urs' condition deteriorates slowly. He needs regular transfusions of platelets and does no longer attend school. During the weeks prior to Christmas, he stays at home doing handicrafts when he feels well enough. He dies two days before Christmas. Care is predominantly provided at home. For the last two days of life, Urs is admitted to the oncology ward as he suffers a lot from bleeding; he wishes explicitly to be on the ward where he feels safe. After Urs' death the contact to the family is maintained through condolence cards from the treating physician and the nurses involved, attendance at the funeral, and a visit of condolence two months later.



**Fig. 1** Vignette 1

To summarize, the definition of PPC is broad; key elements of PPC are: enhancement of quality of life, symptom control, psychosocial and spiritual care, and bereavement care.

## Epidemiology

Given the broadness of the above definitions, specifically with respect to when to initiate PPC, studies on epidemiology may help to better characterize children who might benefit from PPC. Approximately 50% of deaths occur in the first year of life and two thirds of infant deaths occur in the neonatal period [27, 46]. In the USA, Feudtner et al. [15] studied mortality data of children between 1999 and 2006. Three important aspects of death in childhood emerged: (1) infant deaths are mostly caused by perinatal conditions (prematurity, congenital malformations or syndromes, or chromosomal disorders); (2) older children are more likely to die from external causes, such as traumatic injury; and (3) complex chronic conditions (CCCs)—such as congenital and chromosomal disorders, cancer, neurodegenerative diseases, cardiac malformations, or cystic fibrosis—constitute the most important group of diseases responsible for disease-related death in childhood. In Feudtner et al.'s study [15], CCCs were characterized by a duration of at least 12 months unless death occurred earlier, and by the requirement of specialty pediatric care (e.g., hospitalization in a tertiary care center). It can be stated that cancer is still the leading disease-related cause of death in children more than 1 year of age [48].

A further investigation by Feudtner and his group [16] included a retrospective cohort study of 678,365 patients (0–21 years) admitted to Pennsylvania hospitals between 1994 and 2001 and monitored for 1 year following discharge. With their study, they aimed to develop a model that had the ability to predict in-hospital and 1-year post-discharge deaths in children. The adjusted odds of death during hospitalization were most strongly associated with patient's age. Infants <1 month at the time of admission had a 30 times higher likelihood of passing away than children >1 year. With respect to specific types of CCCs, malignancies and metabolic diseases had the highest adjusted odds. In addition, a history of three or more hospitalizations prior to the index hospitalization was associated with an increased risk of death during the hospitalization. During the year following the index hospitalization, the number of prior admissions ( $\geq 3$ ) increased the odds of death 20-fold (95% CI, 15.9–25.7).

In short, 50% of children with serious illnesses die during their first year of life. Causes of death are age-dependent; older children predominantly die from external causes, followed by children with CCCs. Children with CCCs and frequent hospitalizations have a higher risk of dying following a further admission or thereafter.

## The concept of PPC

The core of the concept is defined by the child *suffering from*, but also *living with* and *living despite of* a life-limiting illness. The immediate context for a child will most often be his or her biological family, but it may also be a foster family, and depending on age and developmental condition, this context may widen to encompass additional people such as friends. Therefore, further aspects such as involving close friends in the EOL care at home as well as in the process of leave-taking should be considered. To provide high-quality care that strives to enhance the quality of life of these children and their families, the full range of clinical and educational resources of PPC must be made available [1]. In addition, with respect to adequate care, it is important to define which children could benefit, and at what point of an illness trajectory. Moreover, it needs to be established where and how such care should be provided.

### PPC for whom and when?

As described above, children diagnosed with CCCs such as metabolic diseases or neurodegenerative conditions, with three or more hospitalizations prior to a current admission, may have a significantly higher risk of death [16]. During an in-hospital stay of such a child, a careful and comprehensive discussion with the family is necessary to assess the child's current situation at home. Moreover, such a discussion can initiate the process of reflection on further goals of treatment and care (Vignette 2; Fig. 2).

#### Vignette 2 Progressive incurable neurological disease (ACT group 3 [19])

Caroline is a 4-year-old child with progressive degenerative encephalopathy, suffering from severe tetraplegic cerebral palsy and recurrent seizures. She does not communicate verbally, is wheelchair-bound and nutrition is provided through a gastric tube. On a very low baseline, she seems to enjoy life with her two younger brothers.

For 3 months, Caroline's health deteriorates following a severe pneumonia from which she does not fully recover. Seizures cannot be sufficiently controlled at home, nutrition cannot be provided as before and consequently, the parents have to take her to the emergency room repeatedly.

Two scenarios are conceivable and desirable:

#### 1) Parents expressed their wishes in the case of deterioration

At the very beginning of Caroline's disease, her family decided that they wanted to prevent Caroline from unnecessary suffering and allow natural death to occur when the time for it comes. Upon re-evaluation of her situation by the primary care team, treatment of seizures is modified, nutrition is no longer forced, and outpatient nurses further support home-care and the anticipation of Caroline's death. Caroline dies peacefully at home two months later. The care providers stay in contact with the family during the first months of grief.

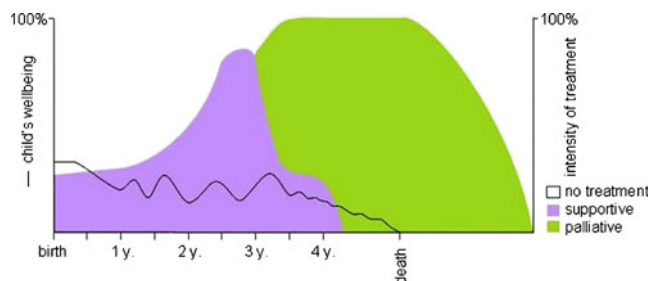


Fig. 2 Vignette 2

#### 2) Parents need some more time

Caroline is admitted to the ward, a work-up is initiated (encephalography, ultrasound of the abdomen) and the nurses who know Caroline observe her for three days. Following this period, a round-table discussion is planned. The nurses confirm what the parents recognized at home and with the team, the family can now decide to take Caroline home and focus on her comfort, expecting that she will soon die. During this time and beyond Caroline's death the family is supported by the outpatient nurses and the pediatrician.

A different, broader and more principled approach centered around four disease categories has been defined by ACT (Table 1 [19]). This approach helps to single out those children with specific diagnoses and their families for whom it would be beneficial to initiate such a discussion, irrespective of disease stage and additional events. The advantage of such an approach may be that the discussion takes place at a time when the child is in a stable condition and the issue of talking about goals of treatment is less threatening to the family. For patients in category 1, particularly children with cancer, PPC is often initiated when relapses or disease progressions are no longer responsive to cancer treatment (Vignette 3). For patients in categories 3 and 4, on the other hand, PPC frequently comes too late in the course of a disease, which may be due to the slow and relatively “uneventful” deterioration of the disease [20]. At any rate, in daily practice, the leading cause for contacting a PPC team is not the diagnosis, but other needs (respite, coordination of community services, physical symptom management, and psychological, emotional, or spiritual support for the family) [20].

Besides an increasing frequency of hospital admissions [16], further triggers and events have been described that should evoke a reflection on treatment goals in a family or within a team. These include episodes of illness without subsequent recovery to the child's usual best level of health, which may also mean gradual loss of function, loss of “normality” [32, 56], or increase of technical or medical support needs. The focus in such cases is shifted from the diagnosis to the physical, psychological, and social impact of the disease [41, 51] and the specific needs of a patient, amounting to a “needs-based definition of palliative care”.



**Table 1** Association for Children's Palliative Care (ACT) Categories [19]

Category	Examples
Group 1 Life-threatening conditions for which curative treatment may be feasible but can fail.	Cancer, heart defects, irreversible organ failures
Group 2 Conditions where premature death is inevitable. Treatment may aim at prolonging life and allowing normal activities.	Cystic fibrosis, Duchenne muscular dystrophy
Group 3 Progressive conditions without curative treatment options. Treatment exclusively palliative, may extend over many years.	Metabolic disorders, neuromuscular diseases
Group 4 Irreversible but non-progressive conditions causing severe disabilities leading to susceptibility to health complications and likelihood of premature death.	Severe cerebral palsy

**Vignette 3** Relapsed nephroblastoma (ACT group 1)

Martin is a 7-year-old boy who has suffered a second relapse of stage IV nephroblastoma. The relapse is diagnosed on the occasion of a routine control checkup 3 months after Martin received autologous stem cell transplantation. The relapse involves the abdomen (including the liver) and lungs. He is otherwise in an excellent condition and just started school. His parents are shocked; in their view, Martin has recovered so well that this information is nearly unbelievable. Accepting that Martin will not survive but die prior to his next birthday, they want to try something new and ask for a treatment that would make it possible for him to attend school and that would allow them as a family to have some more time with him. Oral chemotherapy, which should not lead to myelosuppression, is initiated. When Martin's health begins to deteriorate under palliative chemotherapy 4 months later, developing generalized pain, chemotherapy is stopped and medication to control pain and severe constipation is intensified. The family receives intensive support at home where Martin dies among his beloved parents and sister. The pediatrician who lives nearby the family

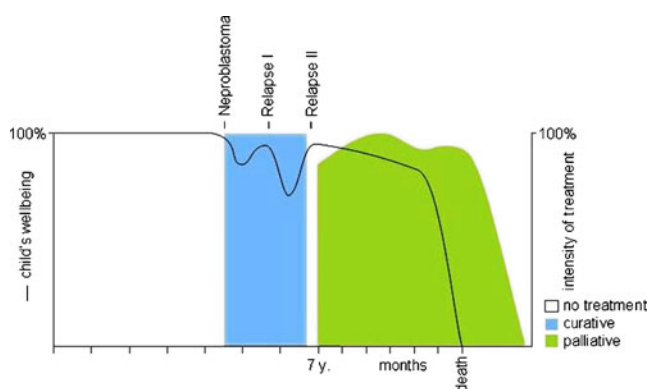
keeps up contact with the family and provides support for Martin's sister. The oncologist visits the family two months later to discuss aspects of the disease and treatment with the parents and to visit Martin's grave with his sister and mother (Fig. 3).

## Disease trajectories and the meaning of transitions

Disease trajectories can be described by focusing on the course of disease with respect to the condition of the child in relation to time [19, 26], or by including the social, psychological, and spiritual wellbeing or distress of the whole family [45]. During the disease trajectory, children and whole families experience multiple transitions on different levels of body, mind and spirit and include, in the words of Hynson, transitions “from healthy to sick, from person to patient, from home to hospital, from parent to nurse [...] from a worrying symptom complex to a diagnosis, from the diagnosis of a potentially curable illness to an incurable illness, from life to death” [22]. Transitions may be associated with a high amount of uncertainty because they may be abrupt or gradual and predictable or unpredictable. Moreover, within a family, the effects of transitions are often experienced asynchronously, which may result in additional conflicts and suffering.

## Location of care

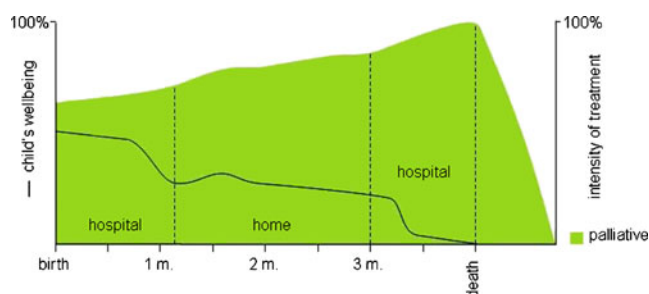
“The unit of care is the child and the family” [23]. To be able to care for a child at home often means a relief for the whole family with regard to organizational balancing between the hospital and the every-day-life at home [23, 30]. However, the home should not be overestimated and equated with “good care,” “good parents,” or “good death.” Just like the family, the

**Fig. 3** Vignette 3

hospital setting, too, may be a “unit of care” where the familiar clinical environment and relationships with staff can offer medical support, security, and social contacts (Vignette 4). In neonates, trauma patients or when unforeseen health deteriorations in children with CCCs occur, appropriate care can only be provided in the hospital and, in some cases, only in intensive care units [4]; in other cases, a more flexible modus of care is feasible. Therefore, it is vital to offer flexible, individualized care, taking into account the particular needs of a child and his or her family [13, 30], and not to be prejudiced towards the location of care.

**Vignette 4** Infant with severe complex syndrome (ACT group 3)

Sarah is a 4-month-old girl with a prenatally diagnosed complex syndrome. She suffers from a complex cardiac malformation with cyanotic spells, dyspnea, severe malnutrition, recurrent aspiration, and episodes of unrest and crying. Following a first diagnostic work-up in her first two weeks of life, she is discharged from the hospital and the family receives support at home from outpatient nurses, the pediatrician and the cardiologist who was involved in the postnatal work-up. After 6 weeks at home, the parents and Sarah's two older siblings seem to be extremely distressed from the care at home and the concurrent attempt to return to the daily routine. Sarah is re-admitted with progressive cardiac failure and recurrent episodes of dyspnea due to a newly acquired aspiration pneumonia. In a multidisciplinary discussion, it is decided to initiate a treatment with the aim of stabilization, including systemic antibiotics and symptomatic treatment of dyspnea with morphine, but to also allow natural death. Sarah has a single room and the family tries to be present whenever possible. Several days later, during the weekend, she dies peacefully with her parents and grandparents present. The nurses as well as the cardiologist will attend her funeral and keep in contact with the family during the following weeks (Fig. 4).



**Fig. 4** Vignette 4

If there is a change in location of care, a key contact person needs to be designated to guarantee not only continuity of care, but also a continuity of communication. Recent studies in Germany and Switzerland have confirmed this need for families to have continuity of care [24, 30]; a pediatric oncologist who was part of the German study, highlighted this need: “They (children) want to remain in the same care team, with the same faces they know and that they have become familiarized to for years” [30]. However, though the need for it is widely acknowledged, continuity of care is not always guaranteed. This has led to the conclusion that (1) in future models for PPC services, the interface between inpatient and outpatient or hospital and home care should provide a high degree of flexibility (including staff working at different sites); and that (2) hospices for children need to be linked efficiently to PPC networks.

### Care provision

The most prevalent model of palliative care provision in acute care hospitals is the consultation service [51]. This model can also be described as shared-care model and has several advantages: (1) the primary attending team stays involved, which allows for continuity of care within the hospital; (2) necessary financial and staff resources may be reduced; (3) PPC knowledge is provided with a clear focus and with the aim of knowledge transfer; and (4) the PPC teams provide their services flexibly and only where needed. The main goals of a palliative care consultation for adult and pediatric patients are to provide specialty-level care for difficult-to-manage symptoms and complex family dynamics, to assist in challenging care decisions, as well as to provide support with logistics or coordination of care [17, 51].

Palliative care is a multi-professional and, if possible, interdisciplinary team approach. This means that the PPC team consists at least of a physician and a nurse and should also include a psychologist and a social worker. Other professionals such as ministers and therapists are further optional team members. Ideally, the PPC team is interdisciplinary and works together with a primary care team of specialists in, for example, neurology or nephrology. Due to the broad and varied knowledge base of its members, such a team can provide adequate patient-centered and family-centered care.

### Practical aspects of PPC in the context of the child and the family

Instead of discussing symptom control, which will only briefly be treated by referencing a very useful guideline,

the focus in what follows will be on frequently encountered issues in day-to-day work with patients and families.

### The meaning of hope

Families of PPC patients wish to receive honest and realistic information, but hope also plays a central role in palliative care. Even though hope is frequently constrained to be a hope of cure, it may also be maintained by shifting the focus to more realistic scenarios. Nevertheless, parents' hope for a miracle may provide stability and does not exclude their realistic acknowledgment of the potential or imminent end of their beloved child's life. In this context, hope may be understood as part of a "healthy denial." Chochinov [7–9], as a result of his interesting investigations into the dignity of terminally ill patients, integrated the concept of hope in his psychotherapeutic interventions for patients near the end of life.

### Communication

Communication is not a specific skill of palliative care; however, it is of particular importance in times of uncertainty and when death is likely to occur [1]. In this situation, many parents wish to receive open and honest information and to be involved in decision-making [42, 43]. Beyond these immediate wishes, communication can have a long-term effect on families which should not be underestimated [44]. Feudtner's comprehensive and compassionate article [14] may serve as a guide to plan a pivotal conversation with a family, to reflect one's personal practice of communication, or to teach communication. His concept of collaborative communication encapsulates both the exchange of information and the collaborative relation between persons who are communicating. It focuses on respect as well as compassion for each other, striving to manage intrapersonal and interpersonal processes that affect how we communicate. For the purposes of this review, one aspect of communication is particularly important, "shifting the perspective." For example, when different treatment options have to be evaluated, some parents tend to persist on their idea of further aggressive treatments. It may, therefore, be necessary and helpful to actively shift the perspective by suggesting to change the vantage point and to assess what the situation looks like from this new perspective. This process may create room and activate new thoughts and prospects. The suffering of the child and the mental picture of a child's death may evoke aversion in parents, particularly when

discussing the "do-not-resuscitate order" which may, for many, conjure up the image of "doing nothing" and simply watching the child die. If a PPC provider succeeds in reaching parents' emotions and contemplating with them what else could be done for the dying child—holding it, lying closely to it, singing, praying—this mental picture of "doing nothing" may gently be changed into a less frightening one of love, closeness, and peace.

### Decision-making

Collaborative and professional communication is a prerequisite for a successful decision-making process. In general, it is recommended to involve children as much as possible and in a developmentally appropriate manner [1]. Adolescents and also chronically ill children are able to take part in conversations and decisions about palliative care. Nevertheless, it needs to be acknowledged that decision-making concerning aspects of end-of-life is exceedingly difficult for parents [31]. The anthropologist Bluebond-Langner has described the challenges implicated in involving children in decisions about care and treatment [3]. On the children's side, several factors need to be considered: (1) their experience with chronic illness plays a major role in their understanding; (2) a differentiated view of illness does not mean that children hold on to the possibility of a cure, through medical or divine intervention; and (3) children may simultaneously hold on to several views of their illness and the efficacy of treatment. On the parents' and professionals' side, three principles need to be considered: (1) decision-making should be conducted without deceit, (2) the participants should be free of coercion, and (3) children have the right to know about the procedures that they undergo. If parents decline to share information with the child, it is important to explore reasons and fears behind this wish and to acknowledge the parents' position. This might help to open the dialog and find a way to get the child involved at least in a small part of the process. It may also be helpful to talk about other parents who have gone through the process of integrating their child in decision-making and who were glad about it [34]. In addition, it may be emphasized that children must be able to trust health professionals and that it is, therefore, essential to be honest with them. In adults, advanced directives are usually in place when end-of-life care is going to start. In children, some kind of a document,



such as a “wish document” [18] could considerably facilitate anticipatory planning of care.

### Anticipatory planning

The ambitious aims of PPC listed in the definitions above may be reached if anticipatory planning including the definition of treatment goals is initiated. To implement such planning, the family needs to pass through the transitions described in the sections on disease trajectories and the process of decision-making. Open, honest, and continuous communication and a high degree of confidence may be helpful; however, in some families this may never be possible or only when death is imminent because it is, on the one hand, an extremely burdensome process for the families, and on the other hand, the process can also be stressful for professionals who try their best and may be disappointed.

Nevertheless, if and when planning takes place, child and family may experience a high amount of quality of life, togetherness, and precious moments that will remain in the memory of those who are left behind [31].

Table 2 provides a list of aspects which should be considered in care planning. Palliative care can be divided into five phases which should be more or less applicable to all children above one year of age, irrespective of their diagnosis: (1) The phase prior to PPC, describes the time when it is recognized that a treatment is no longer effective or the child’s condition slowly begins to deteriorate and a first contact between the primary care team and PPC team is made. (2) The initiation of PPC includes the first contact between the PPC team and the child/family that aims to explain PPC and to develop a holistic care plan. (3) The maintenance phase of PPC should provide some stability for the child as for the family during which the child may enjoy some of the most valuable moments of life—the decision to make a film instead of going to school, or the father’s decision to take off 2 weeks from work and stay with the whole family. (4) The EOL phase is the time when the child might only tolerate certain people present, and symptom control as well as anticipatory grief plays an increasingly important role. (5) The bereavement phase which parents will have been prepared for previously. These aspects are also included in the care pathways developed by ACT [28] and in the “Guidance for discussions about child and family wishes when life is limited” [18].

Symptom management is one part of advanced care planning. Pain and other symptoms have predominantly been studied in children with cancer [33, 37, 54]. To achieve satisfactory symptom control or release, anticipatory

planning and prescribing of measures is mandatory. A very recent and comprehensive manual on the basics of symptom control in palliative care can be highly recommended and is free of charge [29].

In neonatology, the above-mentioned phases will mostly be very short and there is often little time to anticipate. However, it is important that a plan for the phase of bereavement be set in place before the parents leave the hospital.

Older children need to be involved in this process. Children who have experienced illness may have an advanced awareness of illness, dying and death [2]. Kreicbergs et al. [34] found that parents who talked about death and dying with their child did not regret having done so; on the other hand, those parents who did not talk with their child even though they suspected that he or she was aware of imminent death were sorry or even had feelings of guilt.

### The patients’ needs

Older children and adolescents express their needs clearly or give hints which, sometimes, may only be understood in retrospect. Younger children frequently signal their needs in indirect ways, for example by always being present when the doctor comes to discuss serious topics, or by disappearing when a conversation touches upon aspects of their illness.

Adolescents and young adults frequently desire to be involved in EOL discussions [38, 52]. To further explore this desire, the use of an advance care planning document (Five Wishes®) was studied in twenty adolescents and young adults with advanced cancer or a human immunodeficiency virus-1 infection [52]. The “five wishes” address the following aspects: (1) the person I want to make care decisions for me when I cannot, (2) the kind of medical treatment I want or do not want, (3) how comfortable I want to be, (4) how I want people to treat me, and (5) what I want my loved ones to know. The participants of this study found this instrument helpful or very helpful. They were particularly interested in issues of personal comfort and wanted to add more about “who and what they wanted around them at the end-of-life, including visitors, personal items, music and food.” In addition, they suggested more space for elaborating on their wishes. Translated into daily practice, the second wish could, for example, include the assurance to start palliative sedation in an adolescent girl suffering from metastasized osteosarcoma when—in her mind—dyspnea becomes unbearable. It is important, however, to initiate such a discussion when the adolescent is not in a medical crisis [38].

**Table 2** Planning the palliative care approach in the various stages

	Child	Family	Teams
Phase prior to PPC	Who is the child?	Who is the family?	Who are the teams?
	Diagnosis, treatment, prognosis	Parents, siblings, who else belongs to the core family?	Who will be needed for the child at home and in the hospital?
	Physical complaints	Psychosocial situation of the family	Who is the key contact person for the family?
	Knowledge of his or her disease	Knowledge of the disease and prognosis What does the family want and expect?	Diagnosis, treatment, current problems? What different scenarios may occur? What are the plans in these situations and where will they be implemented?
Initiation	Who are you?	Who are you? (in addition to the questions for the child)	Who are we?
	What do you know and what do you need to know?	Are there any discrepancies in understanding or in relationships?	How are we going to work together as a team?
	What are your goals, wishes, and expectations?	What kinds of interventions and support are needed?	How do we provide continuity, security for the child and the family?
	What are your fears with respect to yourself and with respect to your family and friends?	Particular attention needs to be given to the siblings.	Who stays in close contact with the family and gives feedback to the core team?
	What are your needs?	Worst-case scenarios (acute life-threatening event) should be reflected, including resuscitation.	Treatment plan?
	How do you want to be supported now and in the case of becoming unwell?	Preferences for location of care?	Do-not-resuscitate order.
	Where do you want to be treated (home, hospital)?		
Maintenance (re-evaluation and adjustment of treatment and goals)	Living	Being there	Being there
	Hopefully there is a time of peace and space to live autonomously. There may be hope and there may be denial.	Hopefully enjoying moments of togetherness.	Is there need for support, respite?
	There may be wishes for the funeral.		Need for spiritual support, culture-specific requests?
End-of-life	As comfortable as possible to say good-bye.	Being with the child.	Being there and being prepared for interventions Symptom control Prepare the time and moment of death.
Bereavement	Still being a part of the company of grieving family and friends.	There is no rule as to how to grieve and there is no time schedule.	Staying there.

### Parents' needs

Open and honest communication about the onset of palliative care is one of the most frequently cited needs, particularly when death is imminent [37, 39, 44]. However, information, including bad news, should be provided in a most sensitive way, following the principles of collaborative communication described above. Losing a child is an utterly stressful situation for all parents, and they wish to do the

right thing for their child by being “a good parent.” Hinds et al. asked 62 parents of children suffering from cancer to define what being a “good parent” meant to them [21]. Among eight themes, the three most frequent were: (1) “doing right by my child” (89%), defined as making decisions in the child’s best interest and meeting the child’s basic needs to the extent possible, in an unselfish manner; (2) “being there for my child” (48%) in the sense of continuous support; and (3) “conveying love to my child” (42%).

With regard to letting their child go, Kars et al. [31] asked 44 parents of children with cancer in the year before or briefly following death about their “wish of preservation” and “willingness of letting go.” They found that the two aspects coexisted and were linked to the concept of loss. The awareness of death approaching seemed to be a precondition to being able “to let the child go.” All parents experienced a struggle between both feelings; they were able “to let go” with regard to one aspect of care (e.g., chemotherapy) by continuing to preserve another (e.g., special food). Kars et al. [31] concluded that the shift towards letting go was a precondition to taking on the child’s perspective, and that, as a consequence, parents’ awareness of the child’s real situation and needs was able to grow, concurrently with an enhanced ability, “to be there.”

### Siblings’ needs

With respect to palliative care, siblings’ needs and suffering have not yet been studied extensively, but it has been recognized that they need special attention early on in the course of the disease of another child in the family [25]. Because parents are overwhelmed and overburdened by the disease of their ill child, they might overlook the vulnerability of their healthy children who may also need emotional support. Thus, siblings can often benefit from supportive school and camp programs and from professional counseling at the children’s hospital where they can talk about their worries, thoughts, and possible misconceptions. Helpful strategies that can be implemented by parents include dedicating a specific time for the sibling each day or each other day, involving the sibling in the care of the ill child, but also making provisions to allow for “normal life” and distractions to take their usual course. Siblings appreciate to be taken seriously and to be honored for their support, their help in the care for the ill sibling, and their enormous sacrifice.

### Professional caregivers’ needs

The work of palliative care professionals is often considered to be extremely stressful. However, studies have shown that staff working in palliative care suffered less from burnout than other professionals and they experienced job stress levels that were comparable to those of other healthcare professionals working with seriously ill and/or dying persons, particularly clinical oncologists [50]. There is a dearth of literature on how PPC teams care for patients and families and on how they care for themselves and their team members [47]. In their book chapter on team development in PPC teams, Papadatou et al. [47] suggest that teams should develop “holding environments” of similar strength and closeness as those cultivated in families. The concept of a holding environment was developed by Winnicott [53] who used it to describe the role of parents in creating for their infant an environment with safe boundaries which allows them to move from the safety of the

family to the external world. In Papadatou et al.’s opinion [47], creating such an environment for the team is important to cope with the challenges of caring for children with life-limiting illnesses and the repeated encounters with death, and may prevent the depletion of resources in a single professional as well as in a team. The regulation of distress and transformation of suffering shall provide team members with the opportunity to pace their work.

### Bereavement support

Dealing with the death of a child shakes the foundations of a parent’s world but each person will experience such a loss differently [12]. Davies et al. [12] describe grieving as a process of “relearning the world,” an analysis that matches our own experience with parents who went through such a process. Parents may never overcome the death of their loved child, but they will learn to live with and despite this extreme loss. Having this in mind, it is utterly important for caregivers to be available during the first phases of bereavement, to express condolences, and to share moments of grief with these parents [35]. Caregivers are the witnesses of their loss, and to share details of this common experience is often what they seek after the death of their child. Besides these fundamental signs of humanity, bereavement support is extremely important, but still rare in several European countries.

### Future aims

PPC should be accessible for all children and their families who can benefit from it. However, despite all efforts including research to many healthcare professionals not familiar with PPC it is still not clear when to envisage PPC and what this could mean for the individual child and his or her family. Therefore, existing definitions such as the ones of the WHO or ACT require clarification, and criteria of when to implement PPC need to be defined. To meet the needs of individuals but also of health care providers, PPC will require more structures and guidelines besides a high amount of creativity and flexibility.

With respect to research, palliative care for the child as for the adult opens a field of most interesting, innovative and integrative research that has its focus on the needs of the individual and his or her environment. In this context outcome criteria of PPC should receive attention.

### Conclusion

PPC can make a difference in a child’s remaining lifetime by focusing on quality of life and goals that are defined by the child and his or her family. The family is the “unit of care” irrespective of the location of care. To support this “unit of

care,” open and honest communication is essential, particularly when death is imminent. Moreover, bereavement support is an essential part of palliative care, especially in the care of children and their families.

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## Helpful links

Association for Children's Palliative Care (ACT) [www.togetherforshortlives.org.uk/](http://www.togetherforshortlives.org.uk/)  
 European Association for Palliative Care (EAPC) [www.eapcnet.eu/](http://www.eapcnet.eu/)